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CASE REPORT **OLGU SUNUMU**

WEBINO (WALL-EYED BILATERAL INTERNUCLEAR OPHTHALMOPLEGIA) SYNDROME DUE TO MESENCEPHALON INFARCTION

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ABSTRACT

WEBINO (Wall-eyed bilateral internuclear ophthalmoplegia) syndrome is characterized mainly by bilateral internuclear ophthalmoplegia and exotropia. It is caused by lesions affecting the brain stem. In the neuro-ophthalmological examination of an 82-year-old female patient who complained of sudden double vision, bilateral adduction weakness, dissociated nystagmus, exotropia in the primary gaze position, upward gaze limitation, convergence insufficiency findings were observed, and diffusion MRI showed acute mesencephalon infarction. This case, whose clinical findings were compatible with WEBINO syndrome which developed due to ischemic stroke, and was documented with photographs, is presented because of its rarity.
Keywords: WEBINO syndrome, INO, mesencephalon infarction, medial longitudinal fasciculus, oculomotor nucleus.

MEZENSEFALON ENFARKTINA BAĞLI GELİŞEN WEBİNO (WALL-EYED BİLATERAL İNTERNÜKLEER OFTALMOPLEJİ) SENDROMU

ÖZ

WEBİNO (Wall-eyed bilateral internükleer oftalmopleji) sendromu, başlıca bilateral internükleer oftalmopleji ve ekzotropya ile karakterizedir. Beyin sapını etkileyen lezyonlar nedeniyle oluşmaktadır. Ani çift görme şikâyeti ile gelen 82 yaşında kadın hastanın nöro-oftalmolojik muayenesinde bilateral addüksiyon zaafı, disosiye nistagmus, primer bakış pozisyonunda ekzotropya, yukarı bakış kısıtlılığı, konverjans yetmezliği bulguları ve difüzyon MR'ında akut mezensefalon enfarktı izlendi. İskemik inmeye bağlı gelişen, klinik bulguları WEBİNO sendromu ile uyumlu bulunan ve fotoğraflarla dokümante edilen bu olgu nadir görülmesi nedeniyle sunulmuştur.
Anahtar Sözcükler: WEBİNO sendromu, İNO, mezensefalon enfarktı, medial longitudinal fasikulus, okülomotor nükleus.

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INTRODUCTION

WEBINO (Wall-eyed bilateral internuclear ophthalmoplegia) syndrome is characterized by bilateral internuclear ophthalmoplegia (INO) and primary gaze exotropia (1,2). It occurs due to mesencephalon or pontine lesions (1). Mesencephalon lesions may also be accompanied by supranuclear vertical gaze paresis and convergence disorder (3). WEBINO syndrome cases are rare (1). It is known to affect people between the ages of 12 and 85, with a higher incidence in women (4). Considering its etiology, demyelinating diseases, less frequently ischemic cerebrovascular diseases, and other causes were reported (2). Here, a case of WEBINO syndrome, which presented with sudden double vision and developed due to mesencephalon infarction that was documented by cranial MRI and photographs, is presented. Informed consent was signed by the patient and relatives for this report.

CASE REPORT

An eighty-two-year-old female patient was admitted to our emergency department with a complaint of double vision that suddenly started 2 days ago. Her medical history included hypertension, hyperlipidemia, urinary incontinence, ischemic stroke 1 year ago, and hemorrhagic stroke approximately 30 years ago that recovered without sequelae. In her neurological examination, bilateral eyes were deviated outwards in the primary gaze position. When slow pursuit and saccadic eye movements were examined, adduction of both eyes was limited, horizontal nystagmus in the abducting eye with the fast phase in gaze direction and partially restricted upward gaze was observed in both eyes. There was oblique binocular diplopia, and both eyes could not converge (Figure 1). Pupils were isochoric, light reflexes were normal, and other cranial and systemic examinations were evaluated within normal limits. Admission blood pressure was 145/75 mmHg, pulse was 86/minute, and no fever. Sinus rhythm was normal on ECG. Considering the rapidly developing neurological deficit, age, hypertension, hyperlipidemia, and previous stroke history, cerebrovascular disease was first considered. On the diffusion MRI, a bilateral acute ischemic infarction was observed in the dorsomedial aspect of the mesencephalon tegmentum (Figure 2). Cervical MR Angiography

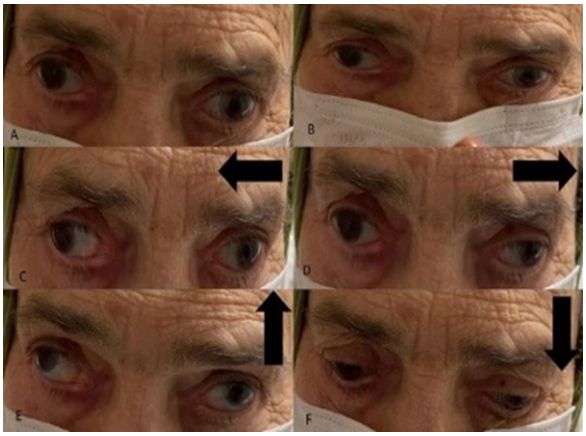

Figure1. Eye movements: A) In the primary position, exotropia in both eyes, B) Convergence insufficiency, C) adduction of the left eye is limited on conjugate gaze to the right, D) adduction of the right eye is limited on left gaze, E) upward gaze of both eyes is partially restricted, F) downward gaze of both eyes is normal (arrows depict the direction of gaze).

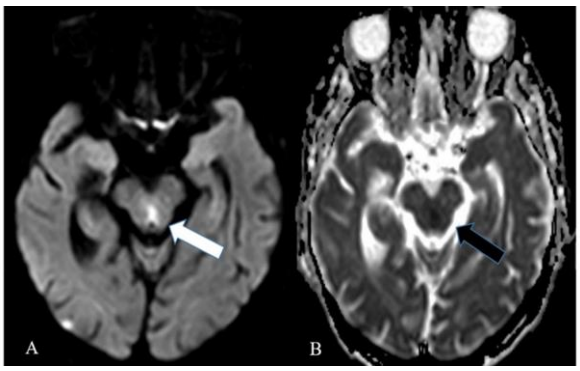

Figure 2. A) Bilateral hyperintensity in the dorsomedial part of the mesencephalon tegmentum in diffusion MRI(white arrow sign),B) In ADC, hypointense acute ischemic stroke was observed (black arrow sign).

(MRA) showed stenosis below 50% in the proximal left internal carotid artery, and hypoplasia of V4 segment of the right vertebral artery and fetal posterior cerebral artery on the left was observed on the cranial MR. The medications she used were amlodipine 10 mg, losartan/hydrochlorothiazide 100/25 mg, atorvastatin 20 mg, acetylsalicylic acid 150 mg, mirabegron 50 mg. In routine tests, LDL cholesterol was 57 mg/dl, triglyceride 165 mg/dl, HDL-cholesterol 41 mg/dl, glucose 97 mg/dl, HgA1c 6%, creatinine 0.96 mg/dl, other liver, kidney, and thyroid function tests and electrolytes were in normal limits. On ECHO, the left atrium diameter was 4.2 cm, left ventricular hypertrophy and wall movements were normal. 24-hour ECG Holter showed no arrhythmia. With the diagnosis

of ischemic cerebrovascular disease, risk factors were evaluated and clopidogrel 75 mg treatment was given. After approximately 3 months of follow-up, diplopia and WEBINO syndrome findings resolved (Figure 3).

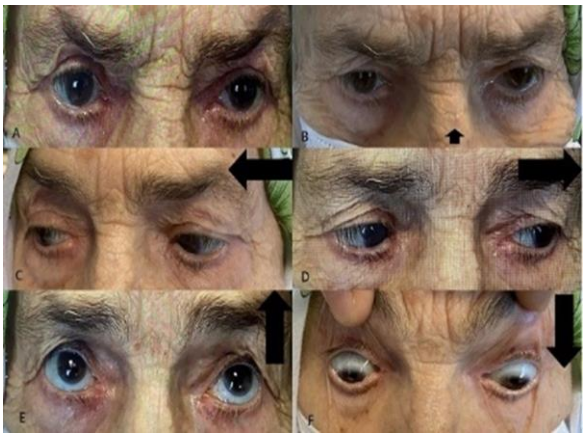

Figure 3. In the follow-up examination performed three months later, eye movements had improved: A) primary position, B) convergence, C) right gaze, D) left gaze, E) upward gaze, F) downward gaze (arrows depict gaze directions).

DISCUSSION AND CONCLUSION

In the neurological examination of the case we present, bilateral adduction weakness and dissociated nystagmus findings indicate bilateral INO. When bilateral INO was evaluated together with findings of exotropia in the primary gaze position, limitation of upward gaze, and convergence insufficiency, it was found to be compatible with WEBINO syndrome.

It is known that bilateral INO develops due to bilateral lesions in the medial longitudinal fasciculus (MLF). MLF is anatomically a pair of white matter fascicles that travel between the interstitial nucleus of Cajal, the rostral interstitial nucleus of the MLF, near the midline within the tegmentum of the midbrain and dorsal pons (2-5). MLF carries signals between the Paramedian Pontine Reticular Formation (PPRF) and cranial nerves 3, 4, and 6 and is mainly involved in extra-ocular muscle movements and vestibulo-ocular reflex (5). Vertical gaze paresis, convergence insufficiency, and sometimes “up-beat” nystagmus are also seen in MLF lesions (4, 6). It is known that the MLF in the midbrain is supplied by the small perforating branches of the P2 segment of the posterior cerebral artery (5).

The pathophysiology of exotropia is not well known and is still debated. It can be seen in both mesencephalon and pontine lesions. It was reported that it develops when the oculomotor nucleus or only the medial rectus subgroup neurons in the mesencephalon are damaged (2). In a study by M. Aktekin et al., it was observed that some subgroup neurons of the oculomotor nucleus, which innervates the medial rectus muscle, were located between the MLF fibers (7). It is thought that exotropia develops when the subgroup neurons of the medial rectus muscle are affected together with MLF due to structural neighbourhood (2). In pontine lesions, the combination of INO and exotropia seen in the contralateral eye is known as Non-Paralytic Pontine Exotropia (NPPE). It was reported that in one case, INO in the right eye and exotropia in the left eye developed due to right pontomesencephalic infarction. It was reported that the cause of this exotropia was the stimulation effect of the contralateral intact PPRF. NPPE is considered to be a mild form of Paralytic Pontine Exotropia (PPE). PPE is characterized by the one-and-a-half syndrome and contralateral exotropia examination findings (8). It is known that the subgroup motor neurons of the medial rectus muscle are also responsible for the convergence function (9).

C-M. Chen suggested that in 3 cases with mesencephalon involvement, a lesion of the medial rectus muscle subgroup neurons was not necessary for exotropia development, and that WEBINO syndrome probably represented the bilateral form of NPPE. However, since MLF axons carry signals from both PPRF and vestibular nuclei, he reported that a pontine lesion was not necessary for WEBINO syndrome to occur and that the MLF lesion was sufficient, and its pathophysiology did not match Wall-eyed monocular internuclear ophthalmoplegia (WEMINO) (1). WEMINO is characterized by unilateral INO and ipsilateral exotropia and was observed in lesions of the pontine tegmentum (10).

During neuro-ophthalmological examination, WEBINO syndrome should be distinguished from WEMINO, PPE, and Non-PPE. In the case we present, bilateral MLF lesion due to mesencephalon infarction explains bilateral INO. The coexistence of bilateral exotropia and convergence insufficiency suggests the involvement of subgroup neurons of the bilateral medial rectus muscle located adjacent to the MLF.

Partial upward gaze limitation was found to be compatible with vertical gaze paresis observed in mesencephalon lesions.

In the etiology of WEBINO syndrome, frequently, demyelinating, and ischemic diseases and, less frequently, degenerative, infectious, inflammatory, toxic, nutritional (e.g., Wernicke encephalopathy, Pernicious anemia), traumatic, post-surgical, neoplastic, metabolic diseases (hepatic encephalopathy, maple syrup disease, Fabri disease) such as hemorrhagic cerebrovascular diseases and progressive supranuclear palsy, and alcohol use and hydrocephalus were reported (2,4,6).

When we look at the literature, spontaneous recoveries were reported in demyelinating diseases, ischemia, and drug-induced WEBINO syndromes. In WEBINO syndrome, treatment is directed against the cause. If diplopia does not resolve, surgical treatments, botulinum toxin injection, prism use, or closure treatments can be applied (2).

In our case, when clinical and neuroimaging findings were evaluated together, acute ischemic cerebrovascular disease was found to be responsible for the etiology of WEBINO syndrome. The patient's diplopia and ophthalmoparesis resolved after 3 months. As WEBINO syndrome is rare and this case is well documented with cranial MRI and photographs, we found it worth presenting.

As a conclusion, ischemic cerebrovascular disease may present with isolated ophthalmoparesis. Clinical recognition of the rare WEBINO syndrome is important in the early diagnosis and treatment of the disease and in the differential diagnosis of other diseases accompanied by ophthalmoparesis

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Ethics

Informed Consent: The authors declared that informed consent form was signed by the patient.

Copyright Transfer Form: Copyright Transfer Form was signed by the authors.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices: GT, SB. Concept: GT, SB. Design: GT, SB. Data Collection or Processing: GT, SB. Analysis or Interpretation: GT, SB. Literature Search: GT, SB. Writing: GT, SB.

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